The Arterial Switch Operation for Transposition of the Great Arteries

Richard A. Jonas

Transposition of the great arteries, together with tetralogy of Fallot, is one of the most common congenital cardiac anomalies resulting in cyanosis. It is hardly surprising, therefore, that shortly after the introduction of cardiopulmonary bypass in the early 1950s, attempts were made at anatomical correction of transposition, ie, the arterial switch procedure. These early attempts were uniformly unsuccessful for a number of reasons:

1. Microvascular techniques were not adequately developed for delicate coronary artery transfer.
2. Pulmonary vascular disease is common in transposition beyond the first year of life.
3. Surgeons failed to appreciate that in the presence of an intact ventricular septum, the left ventricle was inadequately prepared to acutely take over the pressure load of the systemic circulation.

Various atrial level physiological procedures were developed in the late 1950s and early 1960s including the Senning and Mustard procedure. These procedures were widely applied over the next 15 years, particularly after the introduction of balloon atrial septostomy by Rashkind and Miller[1] in 1966. This innovative procedure allowed for a 75% survival rate of transposition patients with an intact ventricular septum. Previously, these patients had either not survived or had to undergo a surgical septectomy procedure, usually the Blalock-Hanlon procedure, which was introduced in 1950.

In 1975, Jatene and associates[2] described the first successful arterial switch procedure in a patient with d-transposition of the great arteries and a large ventricular septal defect (VSD). By this time, thoracic surgeons had become familiar with microvascular techniques necessary for direct surgery on the coronary arteries. Also, there was appreciation of the need for the left ventricle to be adequately prepared.

In 1977, Yacoub and associates[3] described a method to prepare the left ventricle in patients with transposition and intact ventricular septum, thereby potentially extending the arterial switch procedure to a much wider population of patients. However, this approach required an initial palliative procedure followed by an intermediate period of up to several months. In 1983, the concept of primary neonatal arterial switch procedure was introduced by Norwood and Castaneda at Children's Hospital, Boston, MA.[4] This procedure was based on the fact that during fetal life both ventricles are exposed to the same pressure load. Thus, the left ventricle is prepared at birth. However, because pulmonary resistance falls rapidly within 6 to 8 weeks and perhaps as quickly as within 2 to 4 weeks, the left ventricle is no longer prepared. Introduction of an essentially elective complex neonatal operation in 1983 initially met with considerable resistance, particularly because by this time the surgical mortality for the atrial level procedures had reached very low levels. However, a prospective study conducted by the Congenital Heart Surgeons Society[5] showed that the overall survival of an entire population of patients with transposition enrolled in the neonatal period was superior with the neonatal arterial switch approach because of the previously uncaptured deaths that occurred before atrial level procedures that were performed later in infancy. Today, the primary arterial switch procedure in the neonatal period has become the standard of care worldwide.

Indications for the Arterial Switch Procedure

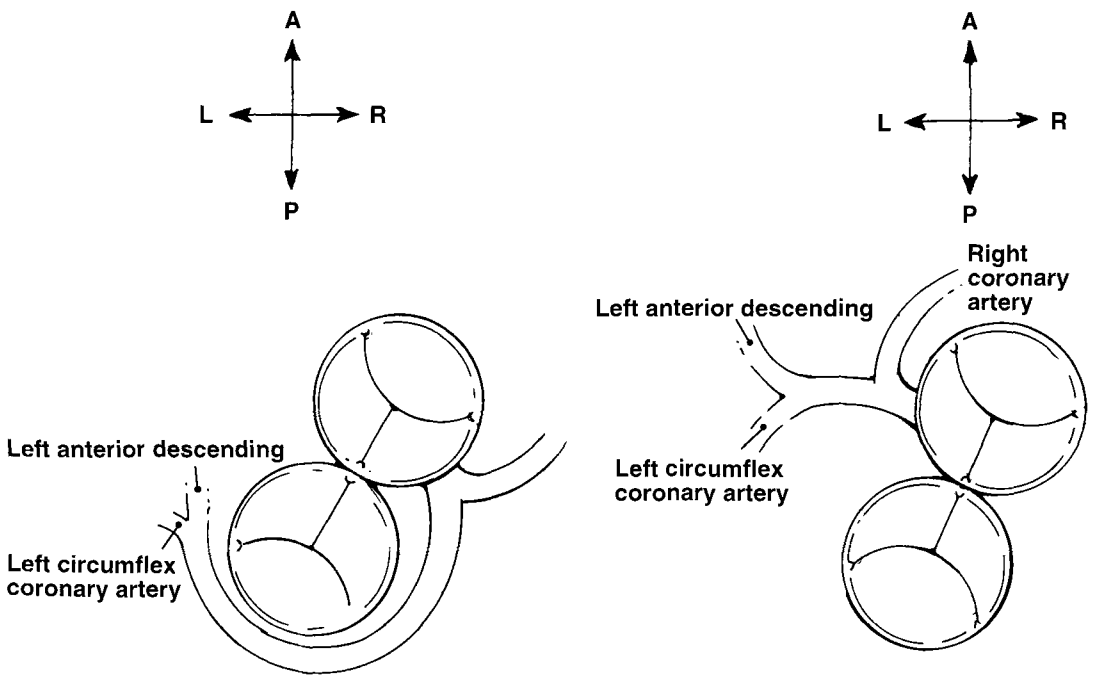
The main population of patients who should undergo the arterial switch procedure are those with d-transposition of the great arteries. In approximately 75% of these patients, the ventricular septum will be intact. A small percentage of patients will have an associated patent ductus arteriosus or secundum atrial septal defect (ASD). We continue to recommend that a balloon atrial septostomy be performed if an adequate sized ASD is not present. The atrial septal defect helps to stabilize the patient during long distance transportation and is also useful intraoperatively in that it facilitates intraoperative decompression of the left heart during cardiopulmonary bypass with a single venous cannula in the right atrium. Patients with transposition and associated VSD or multiple VSDs are also suitable candidates for the arterial switch procedure. Patients with a double-outlet right ventricle and a subpulmonary VSD are generally better suited to an arterial switch procedure than an intraventricular repair. An arterial switch procedure can also be applied in the setting of L-loop transposition of the great arteries (congenitally corrected transposition). In this setting, a concomitant atrial level procedure must also be performed.

Particular Challenges

Coronary Anatomy

Posteriorly placed coronary artery supplying left ventricle. Early in our experience with the arterial switch procedure, we found that coronary artery patterns in which the left ventricle was predominantly supplied by a coronary artery that passed behind the main pulmonary artery from an origin either directly from the right posterior facing sinus or as a branch of the right coronary artery (eg, Figure IA, but not Figure IB) carried a higher risk of myocardial ischemia and death. It appeared that a feedback loop could be set up in which dilation of the left ventricle resulted in tension on this coronary artery that either kinked or flattened the coronary artery and resulted in worse left ventricular ischemia. This then resulted in worse dilation and so on. It was found that accurate placement of the coronary button, particularly with the aid of initial marking sutures placed as the first step of the procedure, as well as careful avoidance of rotation of the aortic anastomosis helped to reduce this risk. Furthermore, with time we tended to mobilize more of the posteriorly placed coronary artery, perhaps as much as 5 or 6 mm, which allowed for some movement of the vessel in response to the changing shape and size of the left ventricle. A subsequent analysis of our more recent experience with the arterial switch procedure has documented that coronary artery anatomy is no longer a risk factor for early mortality (Figure I).[6]

Inverted right and circumflex coronary artery. This coronary pattern is usually seen in association with side by side great arteries with the aorta lying to the right of and perhaps slightly anterior relative to the main pulmonary artery. The right coronary artery and anterior descending coronary artery arise from the anterior facing sinus, whereas the circumflex coronary artery arises from the posterior facing sinus. If the circumflex coronary artery is particularly dominant, the problem as described previously can exist. In addition, because the anterior coronary artery must be transferred in a direct leftward direction because of the side by side nature of the great arteries and because this is directly away from the line of the right coronary artery there is a reasonable probability that the anterior coronary insertion will be under some degree of tension. Although mobilization of the right coronary artery from the atrioventricular (AV) groove can reduce this tension, it is occasionally necessary to extend the anterior coronary button. This can be performed by adding a short tube, usually no more than 5 mm in length, constructed from autologous pericardium.



A Single right coronary artery **B** Single left coronary artery

Fig I. Single right coronary artery (A) previously carried a higher risk of mortality probably because of the long posterior course of the coronary artery supplying the left ventricle. Single left coronary artery (B) does not carry this risk.

Single posterior coronary artery. This is exceedingly rare (Figure II). There is a single ostium arising posteriorly and the right or left coronary artery passes between the aorta and the pulmonary artery. This is a difficult situation to deal with. Probably the best option is to rotate the button 90° posteriorly and to complete the anastomosis of the coronary button to the neoaorta with a hood of autologous pericardium. However, there is significant risk that this area of coronary transfer will be compressed between the neoaorta and the neopulmonary artery. Situations where a single coronary artery arises from either the right or left posterior facing sinus and a coronary artery then runs behind the pulmonary artery can be handled in the usual fashion including the caveat as described previously.

Intramural coronary arteries. We were initially concerned early in our arterial switch experience that the intramural coronary artery represented a high risk situation, but this has not eventuated. Generally, the left coronary artery passes intramurally behind the posterior commissure of the original aortic valve and the ostium emerges in the right posterior facing sinus relatively close to the ostium of the right coronary artery. Although many options are available, we have preferred to excise an intramural segment of the coronary artery on an elongated button that includes the ostium. Even where the two ostia are extremely close together it is possible to split the two coronary arteries into two separate buttons and to transfer these in a relatively routine fashion. It may be necessary to place very fine sutures within the ostium of the coronary arteries themselves. It also is often necessary to detach the posterior commissure of the neopulmonary valve. This can subsequently be resuspended on the pericardial patch, which is used to fill the coronary donor areas.

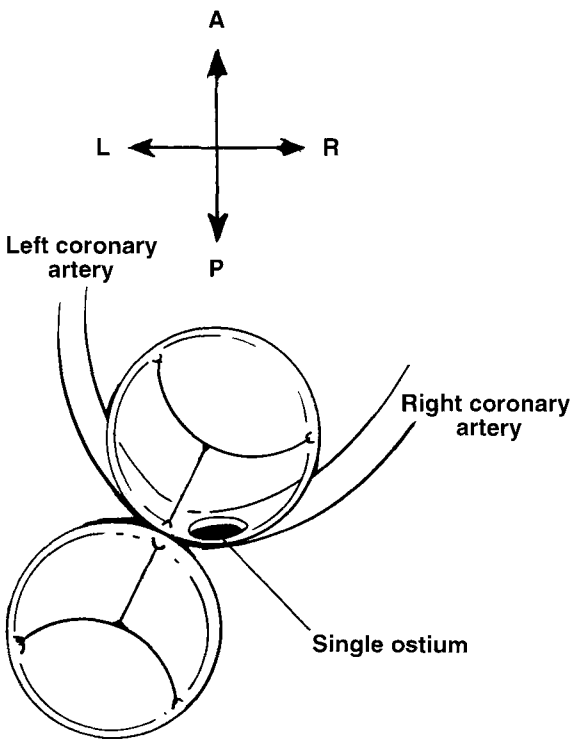


Fig II. Origin of both coronary arteries from a single ostium is exceedingly rare and is a particularly difficult situation to deal with surgically.

Left Ventricular Outflow Tract Obstruction

Left ventricular outflow tract obstruction in association with transposition can be either fixed or dynamic.[7] Fixed left ventricular outflow tract obstruction can be due to hypoplasia of the pulmonary annulus, valvar pulmonary stenosis, fibrous subpulmonary stenosis, or tunnel subpulmonary stenosis. Alternatively, the obstruction may be dynamic, ie, the higher pressure in the right ventricle relative to the left ventricle results in the muscular ventricular septum bulging into the left ventricular outflow tract. Under these circumstances, performance of an arterial switch procedure will result in movement of the septum to the right and may completely alleviate the obstruction. Even in the setting of moderate fixed left-ventricular outflow tract obstruction combined with dynamic left-ventricular outflow tract obstruction it may be preferable to perform an arterial switch procedure relative to the alternative Rastelli procedure. The Rastelli procedure carries with it an obligatory need for reoperation for conduit replacement that often must be performed more frequently than conduit replacement for other lesions because of the immediately retrosternal location of the conduit in transposition. Furthermore, a long baffle pathway from the left ventricle to the aorta in some patients with a Rastelli procedure can lead to late problems of left ventricular outflow tract obstruction.

Age/Left Ventricular Pressure

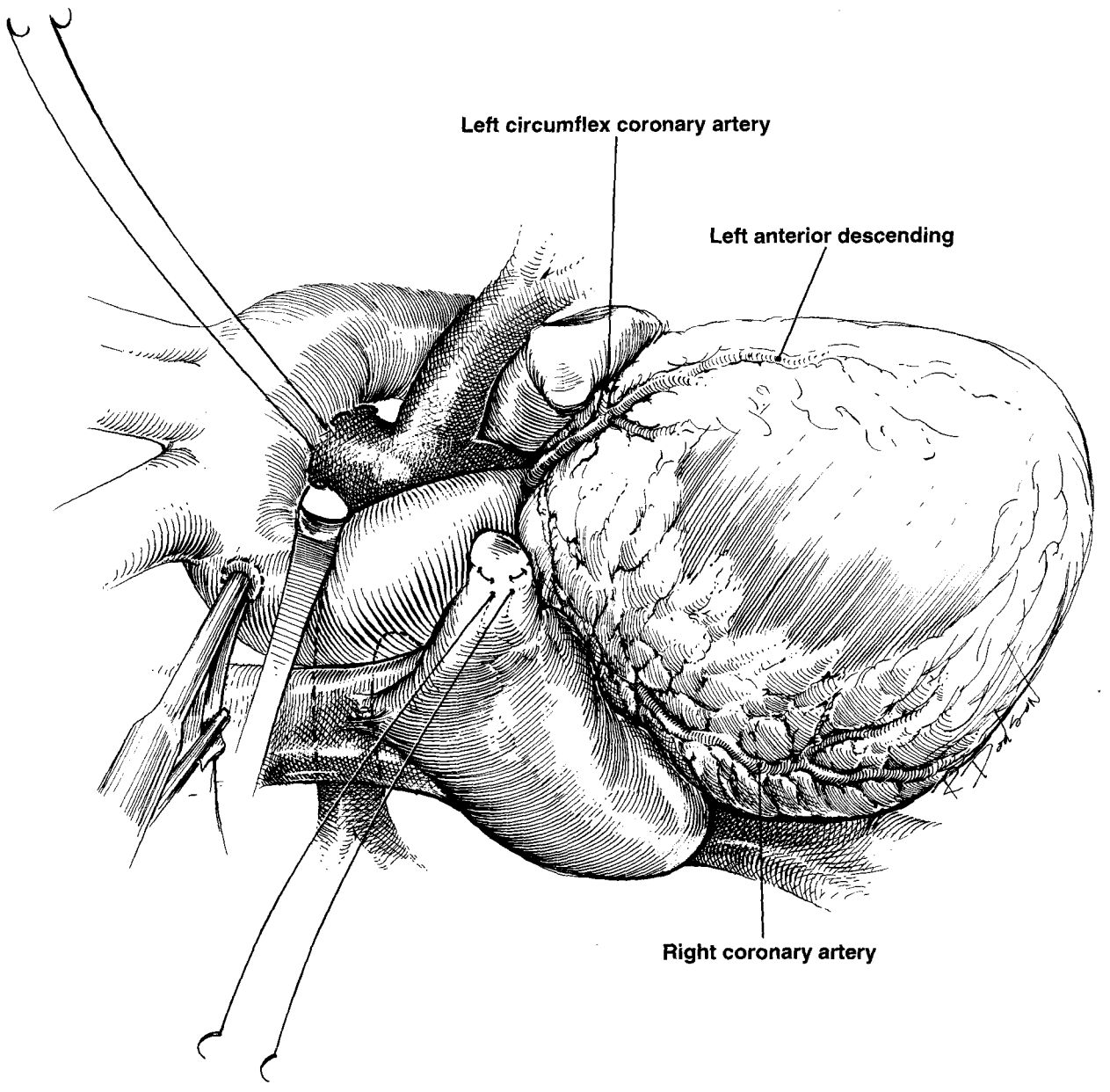
As previously described, the left ventricle loses it ability to acutely take over systemic pressure work because pulmonary resistance falls in the first weeks of life. Some have suggested that it is possible to undertake a primary arterial switch procedure in the young infant with transposition and intact ventricular septum, irrespective of left ventricular pressure, up to approximately 8 weeks of age.[8] Our own practice initially was to undertake an arterial switch procedure as a one-stage procedure in the first month of life and to perform a two-stage procedure with a preliminary banding and shunt in older children whose left ventricular pressure was less than approximately 2/3 of right ventricular

pressure. Although strenuous attempts were made, we were unable to define 2-D echo indices of left ventricular mass and volume that could predict that the left ventricular muscle remained adequately prepared for a one-stage arterial switch.

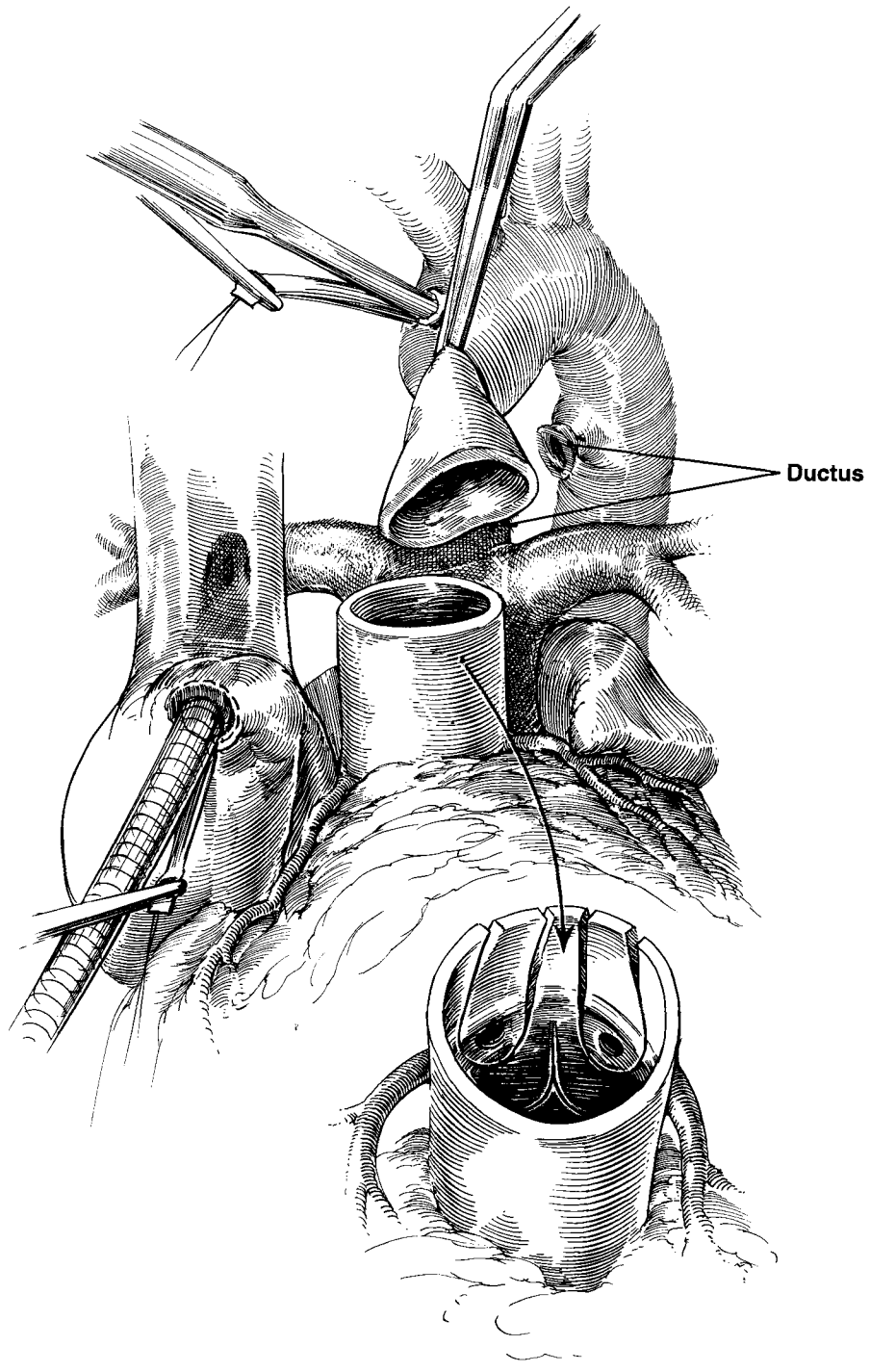
The Congenital Heart Surgeons Society study published in 1988 suggested that the upper limit of safety for a one-stage arterial switch procedure was 2 weeks of age.[5] However, data was collected for this study relatively early in many institutions' experience and may have reflected technical inadequacies at that time. A subsequent analysis performed at the Children's Hospital of Boston failed to show that there was a difference in risk between 1 week and 4 weeks of age.[9] Therefore, our own philosophy in this area is evolving. The approach of performing a primary arterial switch procedure with backup using a ventricular assist device or oxygenator (ECMO) if necessary appears to be not unreasonable in the child who is up to 6 to 8 weeks of age.[8] Fortunately, this question rarely arises in the US, though there are many countries where patients with transposition are first seen beyond the neonatal period and this question is an important one. Beyond 8 weeks of age, most centers would agree that if the left ventricular pressure is less than 60% to 70% of systemic pressure, a two-stage approach should be used.[10] It is our opinion that there is presently no indication for an atrial level repair for d-transposition of the great arteries.

SURGICAL TECHNIQUE

Positioning of the patient is very important. The shoulders must be raised to allow the head to be extended. This tends to draw the great vessels out from the neck. This is particularly important in patients who have a very short neck because they tend to also have short great vessels where the operative field can be quite crowded if the neck is not hyperextended.

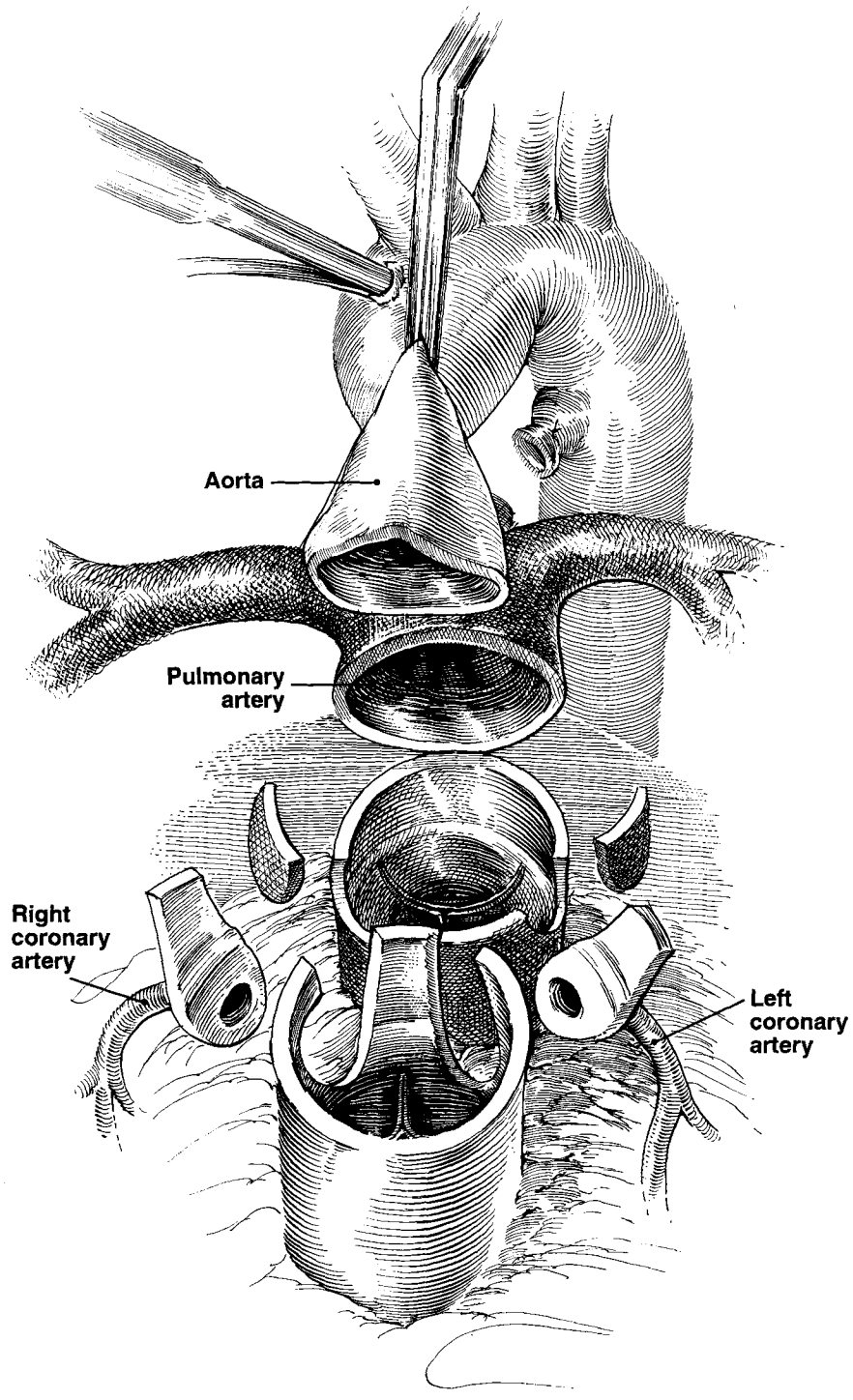


1 Working through a standard median sternotomy, the thymus has been subtotally resected. Marking sutures are placed on the proximal pulmonary artery to indicate the points where the coronaries will be translocated. The sites selected are simply the points at which the artery will most comfortably rotate after mobilization of the first 3 or 4 mm. The arterial cannulation purse-string suture has been placed immediately proximal to the innominate artery. The venous cannulation stitch is placed in the tip of the right atrial appendage. The ductus arteriosus is dissected free and will be doubly suture ligated with 5/0 Prolene (Ethicon, Inc, Somerville, NJ) immediately after commencing bypass with subsequent division.

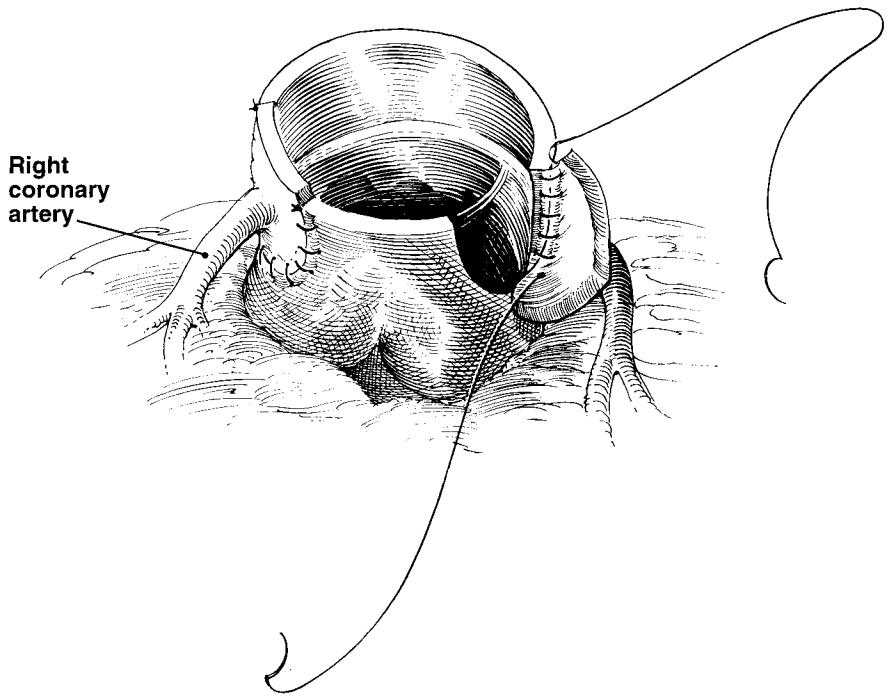


2 The patient is now on cardiopulmonary bypass and has been cooled to a rectal temperature of less than 18°C. A single venous cannula has been placed in the right atrium. The tip of the cannula rests in the orifice of the superior vena cava. The aorta has been cross clamped and a single dose of cardioplegia solution has been infused into the root of the aorta. The aorta has been divided at approximately its midpoint opposite the pulmonary bifurcation.

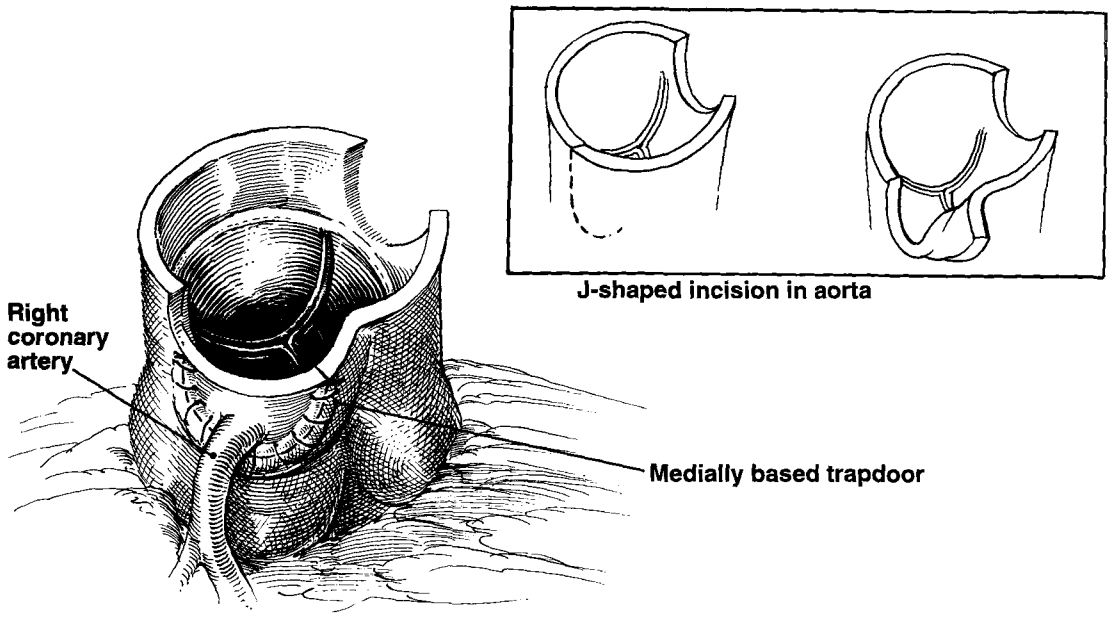
Inset. The coronary arteries are excised with a button composed of the majority of the adjoining sinus of Valsalva. The first 2 to 4 mm of the coronary arteries are mobilized with careful preservation of all branches. If necessary, small epicardial branches are mobilized from under the epicardium.



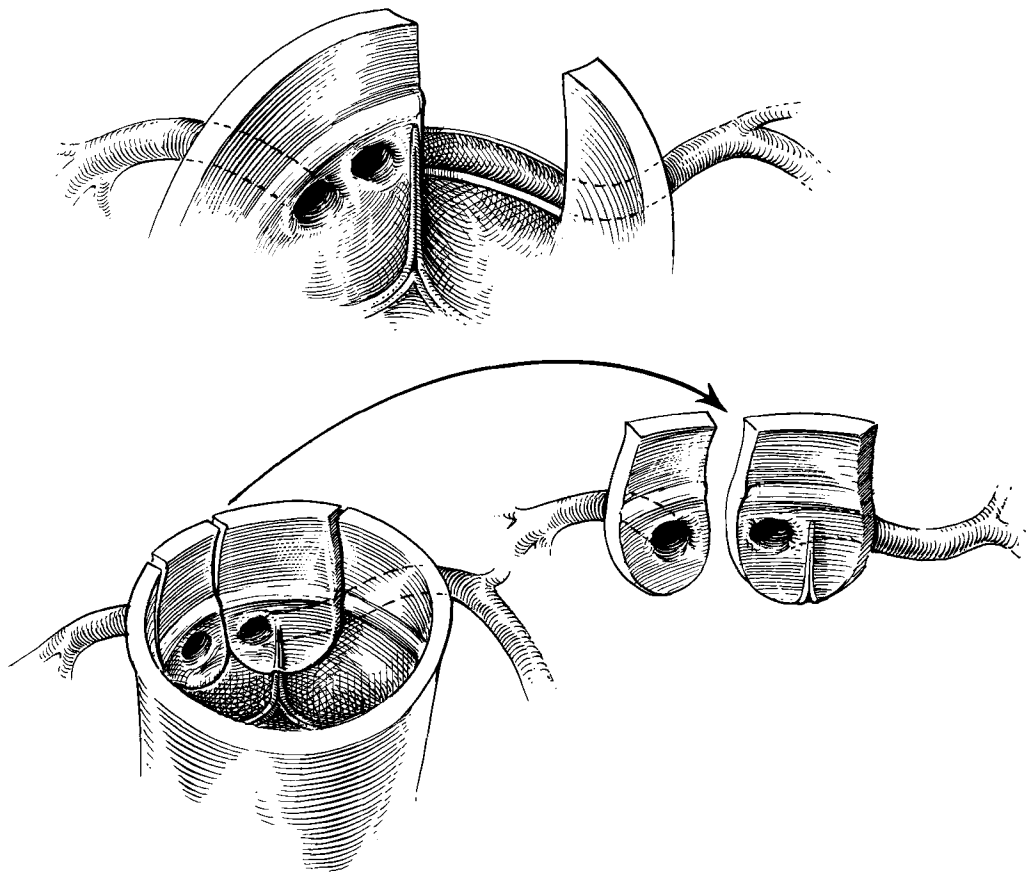
3 A Lecompte maneuver is performed, bringing the pulmonary bifurcation anterior to the ascending aorta. Appropriate U-shaped areas of tissue are excised from the proximal neoaorta. The bottoms of the U-shaped areas are generally at the level of the tops of the commissures of the neoaortic valve. For regular transposition with a subaortic conus and no subpulmonary conus, it should rarely if ever be necessary to excise tissue from below the tops of the commissures of the neoaortic valve. It is important that the original marking sutures be used as a guide to the area of neoaorta that is excised.



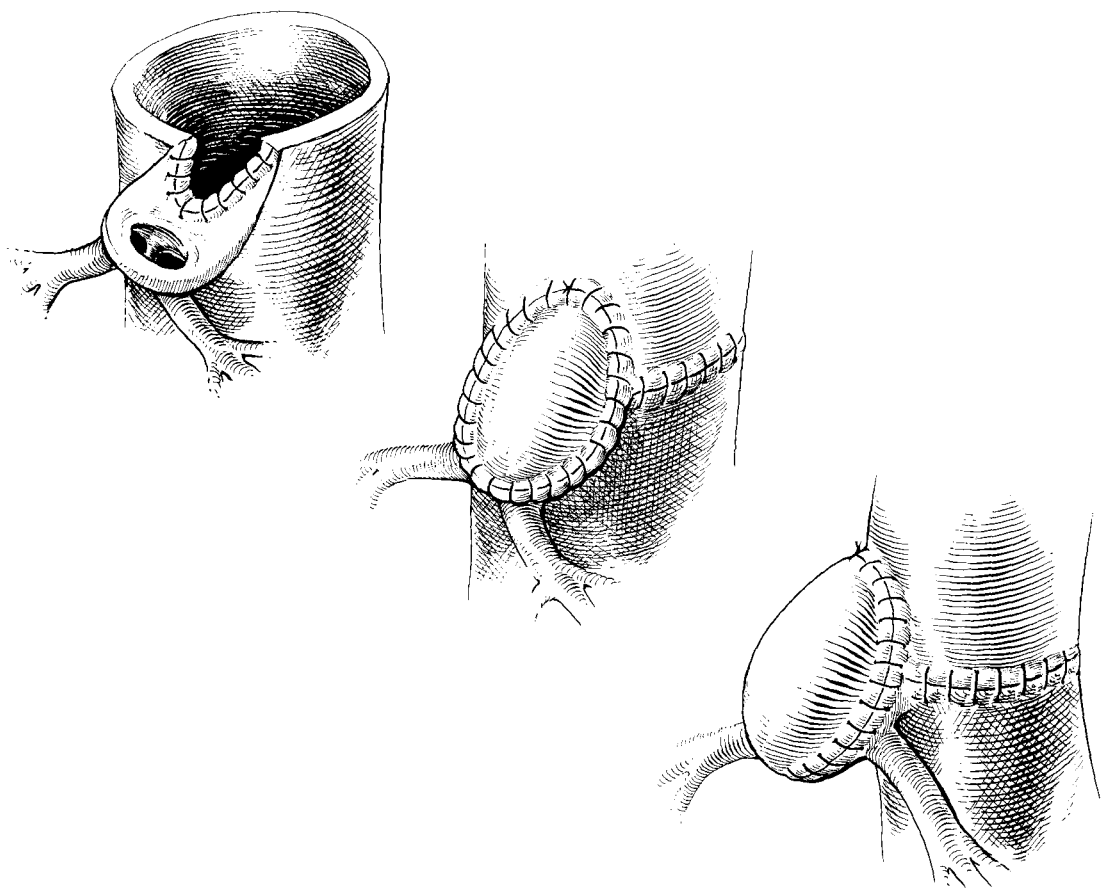
4 The coronary buttons are sutured into the neoaorta using continuous 7/0 Prolene (Ethicon, Inc). The sutures are tied at each end. The suture line is very carefully examined for any very minor imperfections and any suspicious areas are reinforced with interrupted sutures. The areas under the coronary arteries themselves, particularly on the left, are very difficult to expose at the completion of the procedure, therefore, it is critically important that there be no bleeding here.



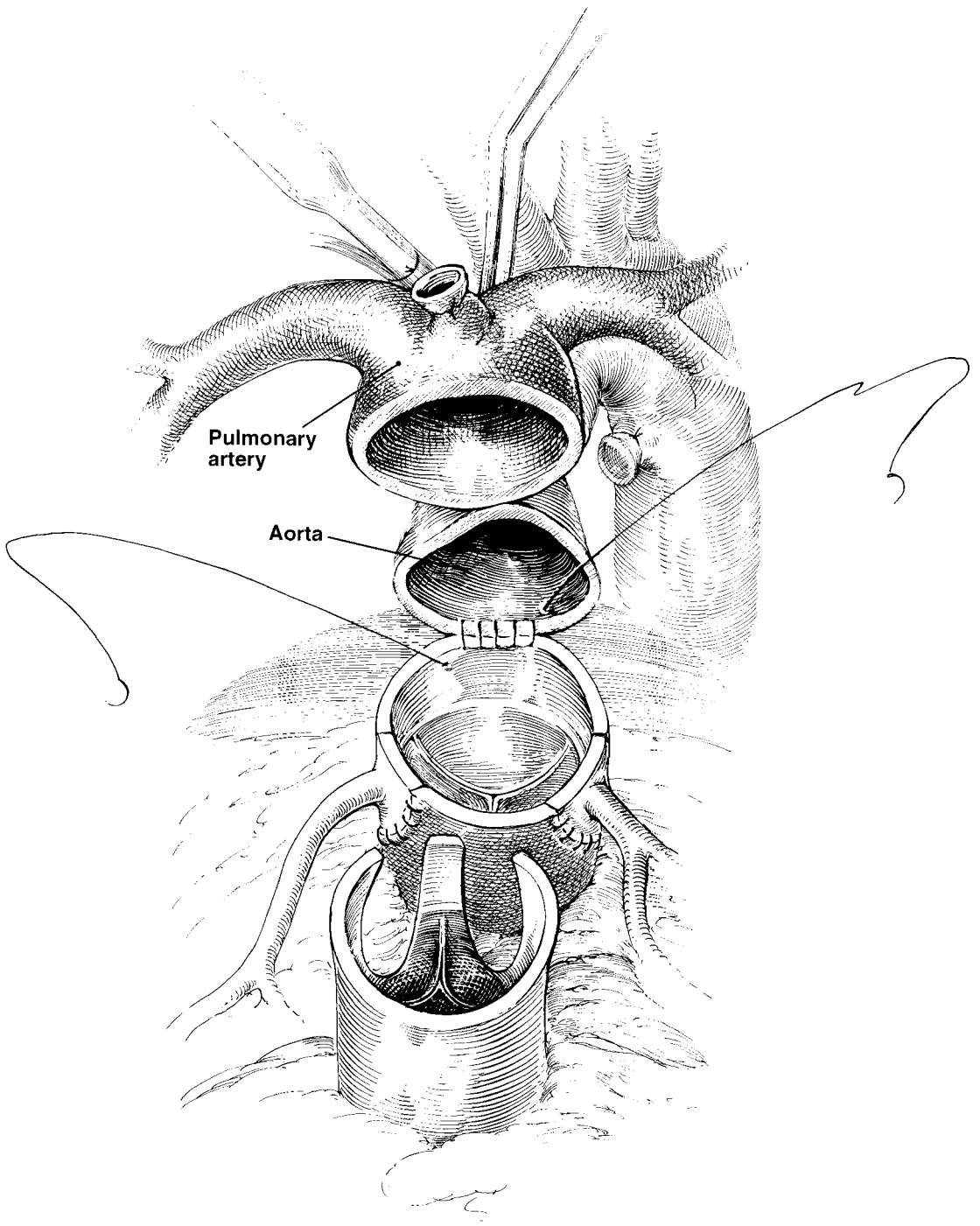
5 An alternative to excising U-shaped areas from the neoaorta is to make a J-shaped incision as indicated in the inset and to rotate a medially based trapdoor. This allows for less rotation of the coronary artery but increases the circumference of the proximal neoaorta, which is generally already somewhat larger than the distal divided ascending aorta. We very rarely find it useful to perform this maneuver. Once again, it can be seen that the coronaries are inserted above the level of the neoaortic valve and, thus, should not interfere long-term with the function of the valve.



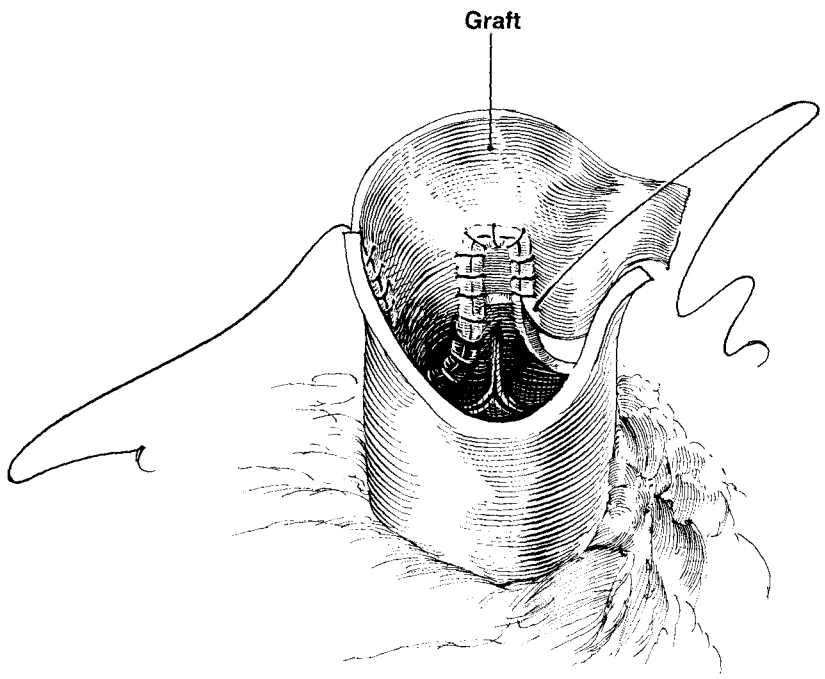
6 If an intramural coronary artery is present, it is usually dealt with by excising a longer button including detachment of the posterior neopulmonary valve commissure if necessary. This commissure can be resuspended when the pulmonary artery has been reconstructed with pericardium.



7 In the rare case of a single coronary artery running between the pulmonary artery and aorta, it is not possible to rotate the coronary button through 180°. The button is rotated through approximately 90° and is then roofed with pericardium.

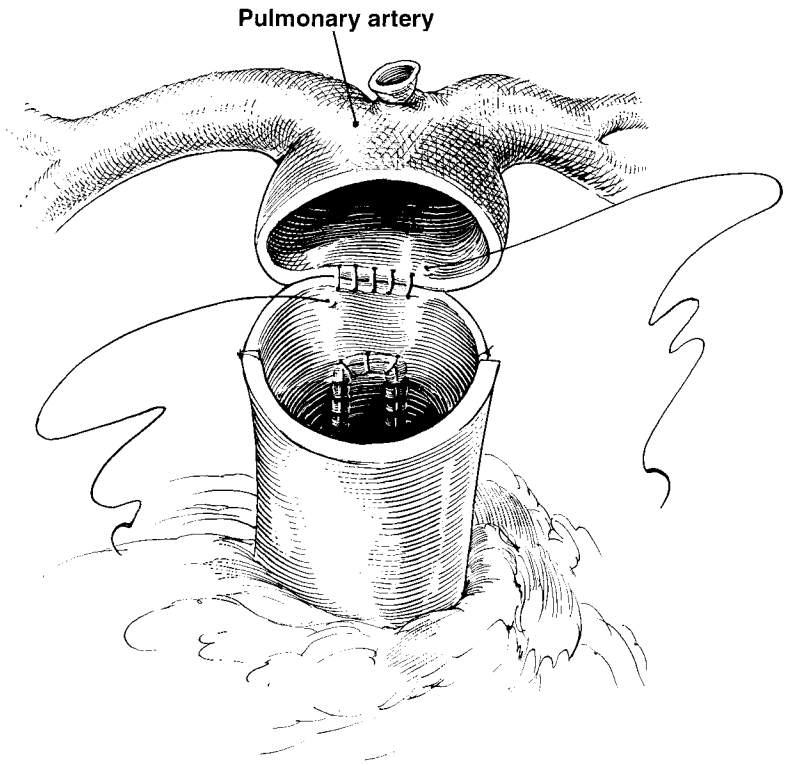


8 The aortic anastomosis is fashioned using continuous 6/0 Prolene (Ethicon, Inc). A continuous suture technique is employed. The areas of junction with the coronary suture lines are reinforced with mattress sutures.

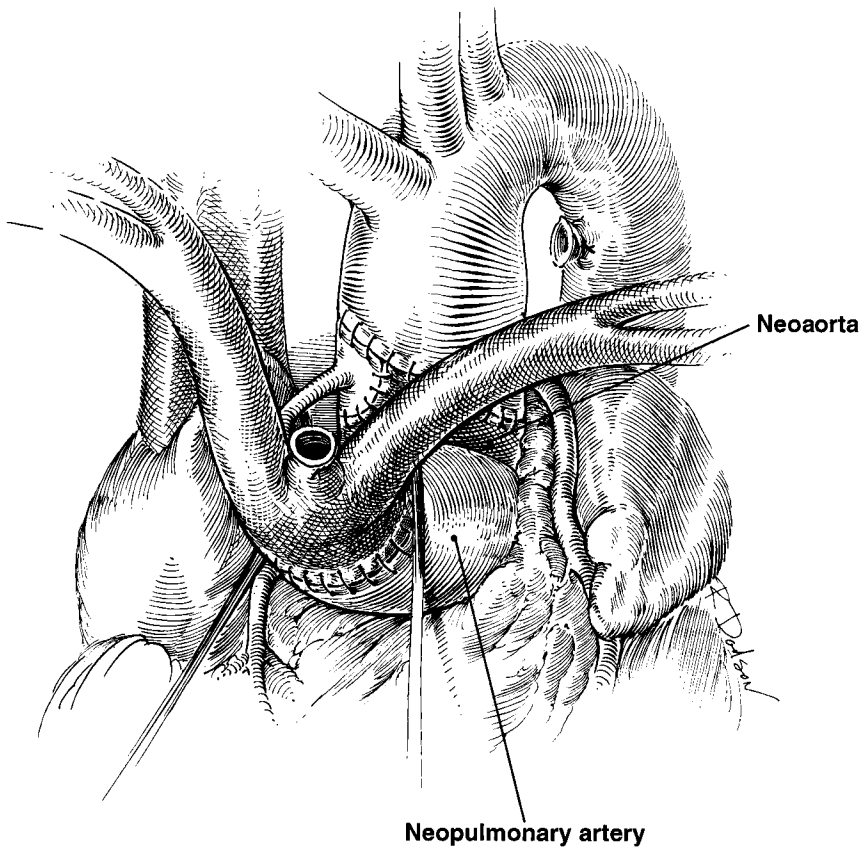


9 The proximal neopulmonary artery is reconstructed with a patch of autologous pericardium treated with 0.6% glutaraldehyde for at least 20 minutes. Continuous 6/0 Prolene (Ethicon, Inc) is employed.

Not shown. The circulation is briefly arrested, though the cannulae are not removed. A short oblique incision is made low in the right-atrial free wall. The ASD is closed by direct suture with continuous 5/0 Prolene (Ethicon, Inc).



10 The aortic cross clamp is released. Satisfactory perfusion of all areas should be observed. The pulmonary anastomosis is fashioned using continuous 6/0 Prolene (Ethicon, Inc). When the posterior layer has been completed, rewarming is begun.



11 During rewarming, a left atrial monitoring line is inserted through the right superior pulmonary vein. Two atrial and one ventricular pacing wire are inserted. With rewarming completed, the child should wean from bypass with dopamine support at 5 μg/kg per minute. After removal of the cannulae, protamine is given. Hemostasis is assisted with thrombin-soaked gelfoam. A right atrial line is inserted through the right atrial appendage. Chest tubes are inserted. The chest is closed with interrupted stainless steel wires to the sternum with continuous Vicryl (Ethicon, Inc) to the presternal fascia and subcutaneous and subcuticular Vicryl, completing wound closure.

Postoperative Care

Monitoring catheters should have been placed in the right and left atrium. In addition, two atrial pacing wires and one ventricular pacing wire also should be available. Intensive care management should be routine. Generally, a low dose dopamine infusion (3 to 5 μg/kg/min) is used, often in combination with milrinone, for afterload reduction. Although in the past it was our policy to heavily sedate neonates for the first 18 to 24 hours postoperatively, this would only be done presently in a neonate who was hemodynamically unstable or who had ongoing bleeding. When it is clear that bleeding is not a problem, weaning from the ventilator is begun. It is usually possible to extubate the child within 48 hours and, frequently today, within 18 to 24 hours of returning to the intensive care unit. It should be exceedingly rare for new problems to develop in the intensive care unit, such as new evidence of myocardial ischemia related to a problem with coronary transfer. This problem should have been suspected and corrected in the operating room prior to transfer to the intensive care unit. The use of thermodilution cardiac output monitors placed in the pulmonary artery in several hundred neonates and young infants after the arterial switch procedure has documented that there is frequently a 10% to 20% decrease in cardiac output between 9 and 18 hours, postoperatively. This is often accompanied by a modest degree of oliguria (1 mL/kg/hr) but other signs of low cardiac output such as diminished peripheral perfusion and hypotension are usually not apparent. Arrhythmias are very uncommon. For example, junctional tachycardia, which is not uncommon after repair of tetralogy of Fallot, is exceedingly rare in patients after the arterial switch procedure, especially when the ventricular septum is intact.

Late Follow-Up

It is important to monitor the growth and development of the main pulmonary artery and branch pulmonary arteries. Failure to perform an adequately large and tension-free anastomosis of the pulmonary artery can result in progressive supravalvar pulmonary stenosis. Annular hypoplasia of the original aortic (neopulmonary) annulus can also result in progressive development of right ventricular outflow tract obstruction. True anastomotic stenosis can sometimes be dealt with by balloon angioplasty, though surgery is indicated if this should become severe. In our experience, coronary artery stenosis and occlusion has been rare and has generally not required intervention because it is usually asymptomatic and related to occlusion of a smaller coronary artery.[9]

In a child who has had left ventricular outflow tract obstruction at the initial procedure, careful follow-up for the development of subaortic or neoaortic valvar stenosis is important.

Results

The reader is referred to numerous reports from Children's Hospital, Boston, MA, documenting the experience with approximately 1,000 patients who have undergone the arterial switch procedure at our institution.[9,11-13] In addition, the report from the Congenital Heart Surgeons Society of the ongoing study of patients with transposition should also be consulted.[5] In general, it should be possible today to perform the arterial switch procedure in patients with or without a VSD who do not have associated significant anomalies such as obstruction of the aortic arch or significant prematurity (eg, birth weight less than 2 kg) with an anticipated mortality of less than 2%. This is the mortality that has been achieved in two large prospective clinical trials at Children's Hospital since 1988.[13]

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